

Shave Excision of Facial Tumors in Tuberous Sclerosis

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Tuberous sclerosis can manifest itself by multiple facial nodules affecting primarily the nose, cheeks, chin, and the nasolabial folds. A simple tangential (shave) excision of these facial tumors is believed to be adequate treatment for some patients.

Introduction

Tuberous sclerosis is one of the large groups of neurocutaneous diseases seen by plastic surgeons, dermatologists, and other practitioners because of their cutaneous presentations. In 1880, Bourneville associated these cerebral lesions with those of the face, and in 1890 Pringle described the skin lesions of tuberous sclerosis in detail. Tuberous sclerosis is a systemic disorder characterized by a triad of facial lesions, epilepsy, and mental retardation. There is no sexual predilection and the inheritance pattern is probably autosomal dominant with variable penetrance. The incidence of tuberous sclerosis is one in 20,000. In the past, the facial lesions have been inaccurately called adenoma sebaceum; more recent studies have classified these nodules as angiofibromas that microscopically show interlacing strands of fibroblasts and collagen with numerous small blood vessels. Histopathologically, tuberous sclerosis manifests itself by hyperplasia of ectodermal cells of the skin, brain, heart, kidney, and retina.

The facial distribution of these nodules is quite characteristic and presents in childhood over the cheeks, nasolabial folds, and chin, as well as over the nose and forehead. The most common locations

overlie the nasolabial folds and the chin. Other skin manifestations include hypomelanotic macules, subungual fibromas, and shagreen patches.

Alvarez first reported surgical excision and grafting of a massive tumor involving the right cheek in 1957, Divir and Hirshowitz describe cryosurgery of the facial lesions in 1980, and Mulliken first successfully treated these tumors with dermabrasion and limited excision in 1977.

A review of the literature over the past 15 years reveals a paucity of case reports describing specific treatments. Two cases of facial lesions as they occur in tuberous sclerosis are presented, and we will describe and discuss the rationale for our mode of therapy.

Case Report 1

LK, a 19-year-old Caucasian man, was referred for treatment of multiple facial tumors. The facial lesions consisted of smooth-surfaced pink to red nodules of various sizes localized to the nasolabial folds, cheeks, chin, and nose. The lesions had been present since the patient was five years old and had enlarged slowly and progressively. The patient also had a history of generalized seizures with mild mental retardation. A cerebral hamartoma was removed at age 15 and a left frontal cyst was drained a few months later. The patient developed hydrocephalus and required a ventricular shunt at 16. Physical examination revealed a well-developed man with mild mental retardation.

Multiple nodules, measuring from 1 mm to 8 mm in diameter, were present on the face (Fig 1). These were particularly abundant over both nasolabial folds and the chin but also involved the nose and the upper lip, with several small nodules present in the left retroauricular region. The patient underwent tangential excision with Nos 10 and 15 blades. The wounds were covered with Xeroform gauze. Within 10 days the skin was healed, allowing easy removal of the gauze. The patient's postoperative appearance two years after surgery is seen in Figure 2.

Case Report 2

SW, an 18-year-old Caucasian man, was admitted for control of seizures and management of skin tumors. The patient had a history of mental retardation and a history of generalized seizure disorder since the age of three months. The facial lesions had been present since he was four. Physical examination showed a well-developed young adult with obvious mild mental retardation. There were multiple nodules present over the nasolabial folds, cheeks, and chin (Fig 3).

This patient underwent tangential excision under general anesthesia, utilizing Nos 10 and 15 scalpel blades. The lesions were excised to the level of adjacent healthy skin. The postoperative treatment was identical to case 1 and good healing took place within 12 days without any complications. Figure 4 is the 12-month follow-up photograph.

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Fig 1.—Preoperative photograph of Case 1.



Fig 2.—Postoperative photograph of Case 1.



Fig 3.—Preoperative photograph of Case 2.

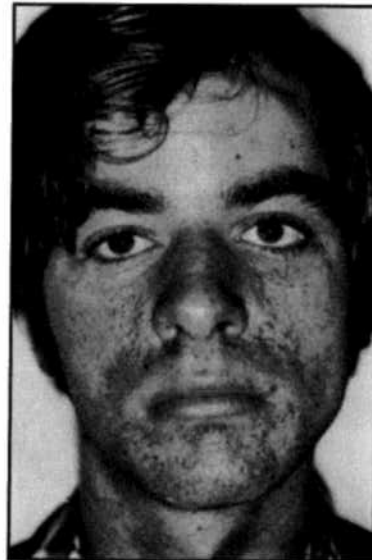


Fig 4.—Postoperative photograph of Case 2.

Discussion

The postoperative results were satisfactory in both cases, and there has been only minimal recurrence of the nodules in a one-year follow-up. We believe this conservative approach is adequate despite the fact that we do not totally eradicate the skin disease. The angiofibromas of tuberous sclerosis involve the full thickness of the skin. In these two cases with extensive involvement, total and complete removal would have required split thickness skin grafting for resurfacing. Such an approach is unwarranted in this clinical setting. These patients do benefit from the aesthetic improvement of subtotal removal, and since these lesions grow slowly, the likelihood that they will require further surgery is small.

In severe cases of tuberous sclerosis, approximately 30% of the patients die before the age of five years and 50% to 75% die before

reaching adulthood. Those patients with lesser signs and symptoms can be expected to live longer, but might require further surgery for recurrent skin tumors—a reasonable approach, considering the minimal morbidity of the procedure recommended.

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